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Case Study of the Month

One-Stage Penectomy and Phalloplasty for Epithelioid Sarcoma of the Penis in an Adolescent

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Abstract

Epithelioid sarcoma (ES) of the penis is a rare tumour. We present a case of ES of the penis in a 16-yr-old boy, for which penectomy and immediate reconstruction with a free forearm phalloplasty, including a urethral reconstruction, was performed. Because total penectomy is a dramatic life event for any patient, the option of immediate penile reconstruction is presented. It can help to prevent major psychological problems after this kind of surgery.

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1. Case report

A 16-yr-old healthy boy was referred for curvature of the penis based on a clinical diagnosis of Peyronie's disease. The boy remembered this curvature for >6 mo, probably longer. He recalled having a lump at the base of the penis for more than a year. Furthermore, he complained of urinary frequency and nocturia. Three months ago he was treated for cystitis, which was not further investigated.

He was unable to describe the exact nature of the curvature. He had observed erections but masturbated only rarely. He described some weakness of the distal part of the penis in erection and deviation to the left side.

On clinical examination an atypical lump was felt at the base of the penis. A 1-cm, hard lesion involving the total circumference of the penile body was palpated (Fig. 1).

On uroflowmetry an obstructive flow pattern was seen. Retrograde urethrography showed a compressing lesion of the urethra that narrowed the lumen.

Magnetic resonance imaging (MRI) revealed a 3-cm irregular rounded mass at the base of the penis. It was contained within both cavernous corpora, compressing the urethra and corpus spongiosum, but without evidence of direct invasion of the latter. The mass showed low signal intensity on T1-weighted images (Fig. 2) and heterogeneously low signal intensity on T2-weighted images (Fig. 3a and b), with inhomogeneous enhancement following intravenous injection

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Fig. 1 – A hard lesion involving the total circumference of the penile body.

of 0.1 mmol/kg gadopentetate dimeglumine (Magnevist®, Shering, Germany; Fig. 4a–d). Distal to the mass, the penile body only poorly enhanced, suggesting severely disturbed blood flow. Cystoscopy showed a white tumour-like structure in the urethra,

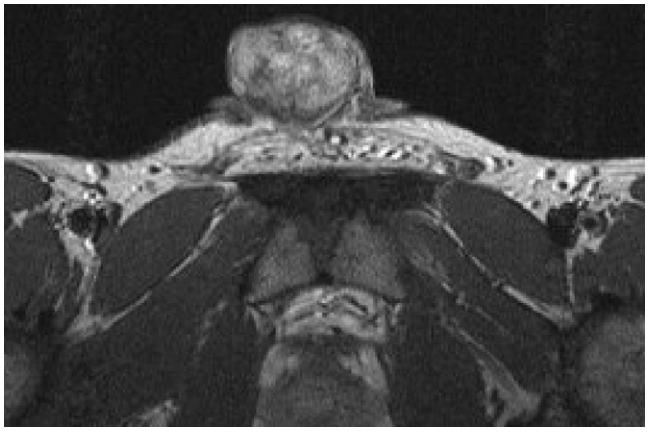

Fig. 2 – Transverse T1-weighted image at the base of the penis showing a mass with low signal intensity disrupting the normal corporeal anatomy.

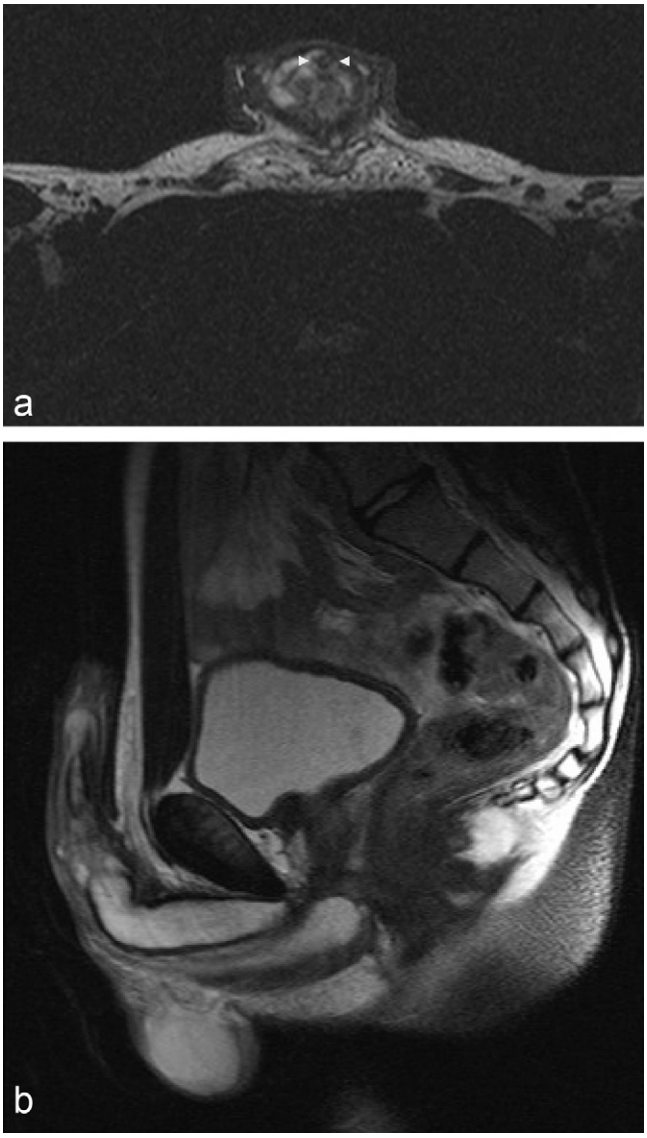

Fig. 3 – Transverse (a) and sagittal (b) T2-weighted images showing an inhomogeneous mass with low signal intensity disrupting both cavernous corpora and compressing the corpus spongiosum and urethra (arrowheads).

filling >80% of the lumen. An excisional biopsy was done through a penile skin incision.

Pathologic examination revealed ES. Metastatic evaluation, including physical examination and computed tomography (CT) of the pelvis, abdomen, and chest, was negative.

After multidisciplinary discussions with pediatric and adult oncology and radiotherapy consultants, it was recommended that surgery be performed at once without any neoadjuvant therapy. It was decided that penectomy was needed. Having a large experience with penile reconstruction in female to male transsexuals [1–4] and in boys without a penis,

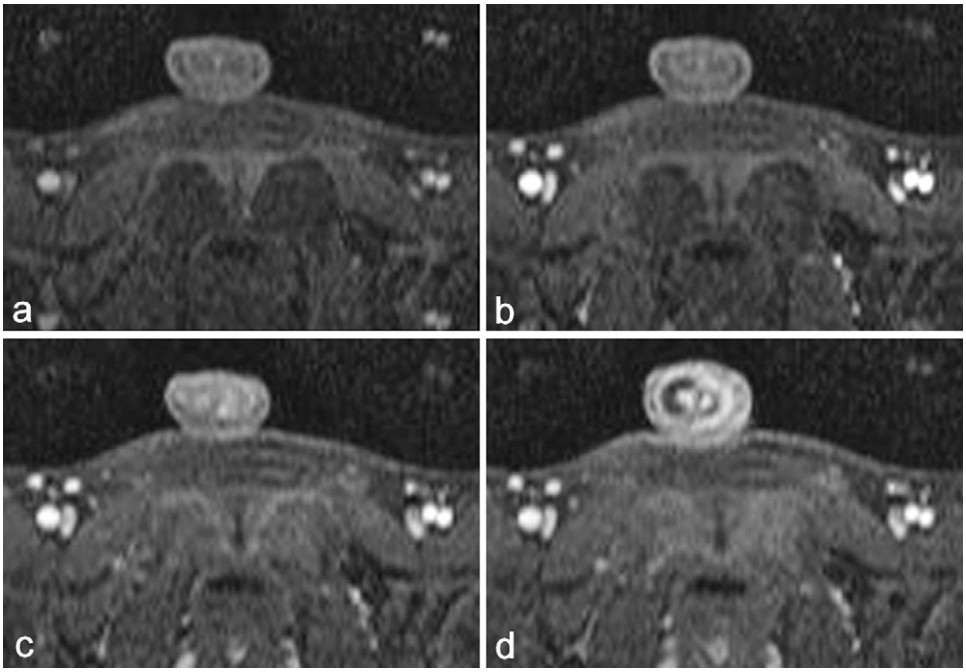

Fig. 4 – Transverse fat-suppressed gradient-echo T1-weighted images before (a) and 30 s (b), 90 s (c), and 150 s (d) after intravenous injection of contrast material, showing peripheral enhancement of the tumour and delayed central enhancement via the central penile arteries.

we discussed this possibility with the patient and his parents. A child psychologist was involved in the discussion. Finally, it was decided to perform the penectomy and the reconstruction in one stage.

A total penectomy was performed. Because the lesion was at the base of the penis, we needed to remove the whole penile body to remove the tumour. The neurovascular bundle was identified and three dorsal nerves could be identified. The cavernous bodies were transected just below the pubic bone and ligated. The urethra was transected and the spongiose tissue was oversewn.

At the same time, the team of plastic surgeons prepared the radial forearm flap and reconstructed a phallus in a tube fashion. At the forearm level, three nerves could be identified to connect with the dorsal nerves. The vascular connection was done with the femoral artery and vein.

Microscopic examination of the penectomy specimen showed a tumour with a nodular growth pattern, composed of a mixed proliferation of eosinophilic epithelioid and spindle cells. Focal necrosis was present. The diagnosis of ES was made. There was immunopositivity for both keratin and CD34. The immunohistochemical examination for desmin and epithelial membrane antigen was negative. Perineural invasion was present, but no evidence of lymphovascular invasion was seen.

Fig. 5 – End result after phalloplasty.

Metaphase cytogenetics revealed a normal karyotype. Surgical margins were negative.

Recovery after surgery was uneventful. In addition to surgical follow-up, the patient was also seen by a psychologist. The phallus healed and voiding was started after 12 d. Voiding was normal on uroflowmetry and no fistulae occurred. The patient was discharged from the hospital after 16 d. The final result is aesthetically good and patient satisfaction is high (Fig. 5).

On follow-up we decided that he would be seen every 6 wk for clinical evaluation during the first year, with CT of the pelvis, abdomen, and chest every 3 mo. Furthermore, he was seen also by his psychologist. After 1 yr an erectile device will be implanted.

Note added in proof

At the time of publication of the manuscript, four small suspicious nodules are seen on CT thorax. Local tumour control is perfect; however, there is possible distant tumour spread for which further diagnosis and eventually surgery and chemotherapy will be proposed.

EU-ACME question

Please visit www.eu-acme.org/europeanurology to answer the below EU-ACME question on-line (the EU-ACME credits will be attributed automatically).

The answer will be given in *Case Study of the Month: Part 2*, which will be published in next month's issue of *European Urology*.

Question:

Both Peyronie's disease and epithelioid sarcoma (ES) of the penis can present as nodules in the penis. The major difference between them is that nodules from Peyronie's disease compared to nodules from ES

A. Never grow into the urethra.
B. Are harder.
C. Are painless.
D. Tend to decrease in size with time.

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